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Clinical Proceedings

of the

CHILDREN'S HOSPITAL

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SPECIAL REPORT

REPORT OF THE BLOOD TRANSFUSION SERVICE

Frank J. Murphy, M.D.
Lois Murphy, M.D.
E. Clarence Rice, M.D.

In the twelve month period ending January 1, 1947 the blood bank received 899 blood donations and gave 1,740 blood transfusions. There were 96 reactions or 5.5% as follows:

Type of Reaction	*Number*	*Percentage*
Major or hemolytic	3	0.17%
Questionable hemolytic	2	0.11%
Minor or non specific	85	4.8%
Allergic	6	0.34%

This is in conformity with that of other blood bank reports.

Erf and Jones[1]	3.2%	in	2,889	transfusions
Levine[1]	4.2%	"	522	"
Carlson[1]	6.0%	"	3,388	"
Stewart[2]	13.6%	"	427	"
Diggs[3]	6.7%	"	1,415	"
Klein[4]	6.5%	"	3,154	"

A further breakdown of the type of reactions are as follows:

Fever alone	49
Fever and chills	15
Chills alone	8
Urticaria	6
Transient cyanosis	3
Headache with fever	3
Dyspnea	2
Transient cyanosis and shock	2
Pain in abdomen with fever	1
Jaundice	1
Hematuria (? due to transfusion)	1
Pulmonary edema death	1
Fever shock death	1
Cyanosis dyspnea death	1

There have been three deaths associated with blood transfusions during the past year.

Case 1: A two weeks old premature weighing 4 lbs. 4 oz. with the diagnosis of exfoliative dermatitis received 35 cc. of a 50 cc. transfusion on its thirteenth hospital day. He became suddenly cyanotic with rapid irregular respirations and expired three hours later. Autopsy revealed multiple lung abscesses resembling a fungus infection and cardiac failure with gener-

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alized congestion of all organs. It is considered that the extra blood volume might have been sufficient to precipitate cardiac failure in the presence of pulmonary disease.

Case 2: A three year old white male weighing 26 lbs. with a microcytic hypochromic anemia and a three month "chest condition" beginning with bronchopneumonia. The roentgen studies revealed haziness over both lung fields with some enlargement of the heart. The patient was given 200 cc. of whole citrated blood. Two hours after the transfusion he developed marked respiratory distress with rales and a rapid enlargement of the liver. The child expired three hours after the transfusion. Autopsy revealed congestive heart failure with dilatation of the right side of the heart, pericardial and pleural effusions and marked congestion of the lungs and liver. There was microscopic evidence of chronic pulmonary infection as evidenced by large pigment bearing round cell macrophages which filled the alveoli. The interpretation was that in the presence of pulmonary disease with the possibility of congestive failure already present, the transfusion was sufficient to overtax the circulation and cause death.

Case 3: A fourteen month old white female weighing 18 lbs. was admitted because of a typhoid fever relapse with a bacteremia, positive stool cultures, an agglutination titer of 1:2560 and a spiking fever varying between 103°–105°F. The patient had moderate abdominal distention and edema. On the seventh hospital day the patient was given 175 cc. of concentrated red blood cells. About three quarters of an hour later the child had a fall in temperature to 98.6°F. which then rose precipitously to 106°F. She became cyanotic and dyspneic with a rapid weak pulse and signs of pulmonary edema. The patient expired one and a half hours after the transfusion. Autopsy revealed cardiac failure as evidenced by an enlarged heart and generalized congestion of all organs. In this instance it was considered that the transfusion was sufficient to push a failing circulation into frank circulatory failure. Falkenstein[5] reports that concentrated red blood cells may be given without dilution with saline for anemia cases. In this instance the patient had 7.5 gm. % hemoglobin which is considered a moderate degree of anemia, but the quantity given was enough for a person twice the size.

Hemolytic reactions have been uncommon in our transfusion service. Usually in this type of reaction an error may be found in the blood typing due to a slow clumping or failure to recognize induced hemolysis in the crossmatching. When a reaction is reported to the blood bank the cross match is repeated and checked by the Pathologist or the Resident in Pathology.

The most common reactions have been non-specific or minor reactions which may be due to poorly distilled water, improperly cleansed apparatus used in the performance of the withdrawal or storage and administration

of blood, a degree of hemolysis present in the donor's blood or low temperature of the transfused blood.

It has been noted in our series and observed by other workers [4, 6, 7] that reactions have a tendency to occur in groups within a 24-48 hour period followed by varying intervals of several days of no reaction. The hypothesis is advanced that improper cleansing of equipment may be the cause; however the correlation is not exact.

The administration of even slightly hemolyzed blood will usually result in a transfusion reaction. All blood with doubtful or slight hemolysis should be discarded on an empirical basis.

Experimentally[8] it has been shown that contamination with staphylococci, streptococci, B. subtilis and diphtheroids will cause extremely rapid hemolysis of a marked degree. Bacteria will grow and will produce extensive hemolysis within 24 hours, even though the blood is kept at an ideal temperature (2°–4°C.). E. coli contamination causes the red cell layer to become reddish black or black.

It has been observed[4] that a sensation of chilliness without fever or evidence of destruction of the transfused erythrocytes frequently accompanies the transfusion of cold blood infused at 4°–6°C. Chills without fever do not occur when blood is given at room temperature.

It has been noted that in seven instances where blood has been given to two patients within an interval of several hours duration that both patients had reactions. In one instance three patients had reactions from blood from the same flask. In order to avoid the future possibility of such occurrences a single patient is cross matched with a donor's blood and not until 24 hours following the transfusion is that donor's blood cross matched with another patient. It is our procedure to discard empirically any blood causing a reaction on the basis of possible unknown substances being present. In another sense, however, the correlation does not hold true because there have been several transfusions from the same flask wherein a reaction occurring in one patient causes no untoward effect in another.

Most minor reactions are due to non-specific agents. The incidence of such reactions have ranged from 1 to 3%. Since most of these are preventable an incidence above 5% is a reflection on the methods in use[1].

Allergic reactions have been uncommon and have consisted of mild urticaria in each instance except one which was accompanied with a transient febrile period.

Blood is collected in a suite of rooms set aside for this purpose by especially trained nurse personnel under a physician's supervision. Donors are selected through the use of a questionnaire. Blood is withdrawn into an inverted vacuum flask containing 75 cc. of a 50% glucose, sodium citrate-citric acid mixture. The blood is stored in a refrigerator in the operating

room at a temperature of 36–40° F. Plasma is prepared from blood that has not been used within a two week period after being drawn.

The grouping, serological examinations, Rh determinations and cross matchings of blood are done by the Clinical Laboratory. It has been our custom to do an Rh determination on all patients prior to the first transfusion.

Since the possibility is ever present in an active pediatric institution of having a case of erythroblastosis fetalis admitted at any time, the policy has been followed of having Rh negative type O blood always available in the bank. This frequently necessitates the use of professional donors. The bank had been supported mainly by relatives and friends of patients and also by donor groups from various civic organizations.

Universal type O blood when the agglutinogen titre is below 1:500 has been used frequently whenever the proper type has not been immediately available.

Absolute contraindications for blood transfusions are pulmonary edema and cardiac decompensation. Relative contraindications are pneumonia and bronchitis. Lung abscesses and empyema are not contraindications[1].

It is also important not to overload the circulation by giving too large a transfusion. As evidenced by the cases here reported, an increased blood volume may bring about right heart failure with fatal results.

Since quantities of blood varying between 50–300 cc. are administered, this means that several transfusions may be given from the same flask. Because of this blood has been dispensed through disposable filters and given by the open burette gravity method. In the future, however, it is intended that the closed system method will be used for all intravenous therapy employing disposable infusion sets.

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PRIAPISM DUE TO THROMBOSIS IN SICKLE CELL ANEMIA

Case Report No. 104

Edward Sawan, M.D. H.R. 47–7002

This is the case of a 10 year old colored male who was admitted to Children's Hospital on July 8, 1947 with the chief complaint of a penile erection of 48 hours duration. The erection had developed while he was asleep. He had associated dysuria but no urgency or frequency.

The patient had never been sick prior to this illness. He was born after normal gestation, weighing 7½ lbs. The delivery was normal and the neonatal period was uneventful. He was breast fed for two months. His vitamin intake was adequate. The developmental history was normal. His mother had died of "anemia" while he was still an infant. There were no siblings. To date he has had nocturnal enuresis, but no other genito-urinary symptomatology.

Physical examination on admission revealed a well developed, slender, colored male who did not appear acutely ill. The temperature was 102°F. pulse 100 and respirations 25. His blood pressure was 100/60. Positive physical findings included icteric sclerae, a moderately inflamed pharynx, hypertrophied tonsils and cervical adenopathy. The thorax was symmetrical and the lungs clear. There was a soft non-radiating systolic murmur heard best in the 2nd left interspace. The liver was 4 cm. below the right costal border. It was non-tender, smooth and firm. The penis was erect and edematous. It was tender along its entire course but greatest at its root where there was a bulbous swelling. There were no palpable cords or masses along the shaft. The foreskin retracted easily exposing the glans. There was no apparent urethral discharge or any evidence of trauma. The right inguinal lymph nodes were palpable but not tender. The scrotal sac was not edematous or tender. The testicles were apparently normal.

Admission laboratory work revealed a hemogram of 3,000,000 with 7.5 gms. hemoglobin. The white-cell count was 40,000 with 72% neutrophiles of which 63% were segmented, 4% band and 5% young forms and 23% lymphocytes. Ninety per cent sickling of the red cells was present. There was also anisocytosis, poikilocytosis and achromia. Repeated urinalyses were negative. Icterus index was 70 units dropping in five days to 14 units. The Van den Bergh was qualitatively delayed with 2.4 mg./100 cc. of bilirubin. The prothrombin time was 105% of normal; the cephalin flocculation was 2 plus in 24 hours and 3 plus in 48 hours. The Harrison-Godfried test for bile in the urine was faintly positive. The spinal fluid findings were normal. The blood culture was negative.

X-ray of the chest, lower lumbar spine, pelvis, sacrum and coccyx was interpreted as negative.

The penis became progressively worse with the edema extending to the scrotal sac. The liver was 6 cm. below the right costal margin. After 48 hours the patient began to improve. The liver receded, the penis became smaller and the temperature dropped by lysis. Complete regression of the priapism occurred within 10 days.

The treatment consisted of two blood transfusions and penicillin, 50,000 units every 3 hours for five days. Locally, wet compresses were used. The patient was discharged 14 days after admission.

DISCUSSION

Dr. Robert D. Cawley: Priapism is a condition of persistent and prolonged erection of the penis, unaccompanied by sexual desire, usually painful and not relieved by intercourse. This condition is not common but is not to be confused with prolonged intermittent erections which often occur in diseases of the genito-urinary tract. Persistant priapism is commonly due to spinal cord disease and is often a symptom of leukemia and new growths. Idiopathic priapism is occasionally seen and the etiology of this remains obscure. With the exception of those cases due to either central or peripheral nervous origin, one underlying factor is present in all instances of priapism, namely, a change in the consistency of the blood found in the venous circulation of the penis resulting most probably from the formation of minute thrombi. The etiology in this case was evidently a thrombosis of the corpora cavernosa occurring during a sickle cell crisis. It is a rare complication of sickle cell anemia and represents the only instance of its occurrence seen at this hospital in spite of a comparatively large number of sickle cell anemia cases seen each year. Dawson has reported 2 cases of priapism associated with sickle cell anemia.[1]

The onset of priapism is sudden and may occur at any age. In true priapism there is an erection of only the corpora cavernosa in the vast majority of cases. The glans and corpus spongiosum show no erection. Thrombosis of the corpora cavernosa is said to be present in all cases over 48 hours duration. The blood is not clotted but is viscous with more or less definite formation of thrombi.

Optimal treatment is considered to be the removal of the thickened or thrombosed blood from the veins of the corpora cavernosa as soon as possible; otherwise the patient will become impotent through losing the power of erection. McKay and Colston recommended the use of a No. 8 needle injected into one or the other corpora cavernosa under local anesthesia being sure that the point has penetrated Buck's fascia and that the dorsal vein of the penis has not been perforated. After removal of the

viscus blood, irrigation may be performed with physiological saline solution. This treatment can be repeated as often as required and is effectual even when only small quantities of blood are removed. The absorption of residual thrombi is promoted by hot baths. Sedatives, narcotics, hypnotics, spinal and deep anesthesia have all been tried without success[2]. In the case reported here, conservative supportive therapy was used with subsidence of the priapism within about 10 days.

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SPECIAL REPORT

INFECTIOUS HEPATITIS

A Review of Thirty-Two Cases in Children's Hospital

Martin Randolph, M.D.
Alfred DeVito, M.D.

Infectious hepatitis has in recent years attained great prominence among the roster of diseases. Numerous epidemics and endemics have been observed since Bamberger first described it as "Catarrhal Jaundice" in 1855.

In the recent war years, the British regarded it as "one or the three most important diseases in the Mediterranean theatre"[2]. Although much has been written on the subject, little has pertained to the clinical manifestations of this entity in children. It is considered, therefore, that a summary of the observations of thirty-two cases of infectious hepatitis admitted to this hospital would be worthy of report.

The following account is based on the clinical records of thirty-two patients admitted to the Children's Hospital between January 1st, 1943 and July 1st, 1947.

The average age was eight years, the youngest being eight months and the oldest child 12 years. Females predominated in a ratio of three to one. The great majority of cases occurred in the fall and winter while only two cases were admitted in the spring; no case occurred in the summer. A history of a recent upper respiratory infection was obtained in only two cases and in no case was there a preceding history of enteral infection.

PRE-ICTERIC STAGE

The onset was abrupt in every case with nausea, vomiting, abdominal pain and fever appearing as the cardinal presenting symptoms. Frequent vomiting was present in every case and was described as projectile in some cases. Abdominal pain was a symptom in 50% of our series; it was intermittent in character and was localized to the periumbilical area in those patients old enough to describe their symptoms. Anorexia was not uncommon. Neither diarrhea nor constipation was a conspicuous complaint in any case. The temperature was normal in three cases and ranged between 100° and 102° in the remainder of the series. The highest temperature was observed during the first two days of the preicteric stage, often reverting to normal in 24 hours. Headache was present in only one case; chills and general aches and pains were not observed.

The stools became lighter and the urine darker on an average of one to two days before the onset of jaundice. In many instances this was a helpful sign in arriving at the correct clinical diagnosis.

The above symptoms persisted for four to ten days (average 7 days) before jaundice appeared.

ICTERIC STAGE

The initial symptoms persisted for one to three days after the onset of jaundice. This remission of symptoms was not as striking as described by some authors but rather a gradual subsidence of symptoms with a return of appetite and a feeling of well-being in three or four days. Fever did not exist in the jaundice stage in any patient.

Jaundice varied in degree from a slight scleral tinge to a deep greenish pigmentation of the entire body reaching its maximum intensity in from two to five days. Clinical icterus persisted on an average of 14 days, the duration ranging from two to twenty-four days. There was no definite correlation between the degree of icterus and the severity of the symptoms.

Pruritis was a symptom in only one patient; bradycardia was not observed in any case.

Enlargement of the liver was noted in 60% of our series, the average being 4 centimeters below the costal margin. Right upper quadrant tenderness was present in eight cases and was accompanied by liver enlargement in five cases while in the remaining three cases there was no associated hepatomegaly. Splenic enlargement was noted in only three cases.

LABORATORY DETERMINATIONS

The urine was positive for bile in the late pre-icteric and icteric stage in three-fourths of our cases; the stools were positive for bile in all cases. Qualitative Van den Bergh reactions were uniformly immediate and direct; these did not change with later determinations. Icteric indices ranged from 24 to 126. Cephalin flocculation tests were positive in every case after 48 hours ranging from one plus to three plus with most cases showing a two plus reaction. Prothrombin times averaged 63%.

PROGNOSIS

All the patients recovered and were discharged in apparent good health, there being no mortality in this series. The duration of hospital stay ranged from two to 24 days with an average of 9 days. There were no relapses and no complications during the period of hospitalization. No extensive follow-up study was attempted, however.

DISCUSSION

The accepted belief that infectious hepatitis has as its etiology a virus is in the main supported by transmission experiments[1] and the epidemiological finding that contacts develop the disease approximately 30 days after exposure to a known case[1, 2, 3, 6]. Yet while the disease suggests a virus etiology, with one exception, it has not been possible to transmit the disease to any experimental animal[1]. Attempts at tissue culture have been unsuccessful[2] and the characteristic inclusion bodies have not been described. Hence the virus etiology of infectious hepatitis has yet to be proven unequivocably.

Although tacitly listed as a communicable disease, infectious hepatitis was not respected as such[1] until it assumed epidemic proportions in the Mediterranean theatre during World War II, ranking with malaria in the depletion of fighting strength[1, 2]. It was the subject of much epidemiologic investigation. Fomites, water supply, flies and droplet infection were variously indicted as the mode of spread without any definite conclusions being reached. In 1945 Stokes and Neefe[5], proceeding on the evidence that the agent responsible for infectious hepatitis was present in blood during the pre-icteric and early icteric stages of the disease, reasoned that such neutralizing antibodies present in globulin might be effective in attenuating or aborting the disease if administered during the incubation period or pre-icteric stage. Accordingly during an epidemic of infectious hepatitis in a children's summer camp these workers administered gamma globulin intramuscularly to 53 of 331 persons who showed no evidence of the disease. Their results indicate that gamma globulin is effective in preventing or attenuating infectious hepatitis when given to exposed persons during the incubation period of the disease. These authors indicated that while the best results are obtained with gamma globulin if given early in the incubation period, it may be of therapeutic value if given early in the pre-icteric stage of the disease.

Clinically, our typical case was manifested by the abrupt onset of nausea, vomiting and abdominal pain with fever of 100 to 101° in a previously well child. These symptoms persisted for an average of seven days before jaundice appeared. With the onset of jaundice, there was a gradual remission of symptoms over a one to three day period. Fever was only present in the first few days of the pre-icteric phase and never exceeded 102°. Jaundice reached its maximum intensity in 2 to 5 days and persisted for an average of 14 days.

These clinical findings are in general agreement with those described by Horstmann and Haven[3] reporting on two institutional outbreaks among children in New Haven, by Cameron[1] and Witts[2] reporting on this disease in the military age group and by others[4–6].

Infectious hepatitis largely is a benign disease although progression to a diffuse hepatitis, cirrhosis and death have been reported[1, 3]. The steady increase in the incidence of the disease in recent years, our comparative inability to deal with it prophylactically and the high morbidity serve to emphasize its importance and represents a challenge in the realm of preventive medicine and epidemiology.

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CONGENITAL MULTILOCULAR RENAL CYST IN A NEWBORN INFANT

Case Report No. 105

Martin Randolph, M.D. J. K. P. 47–4895

This patient was born at a local hospital on April 30, 1947 of a normal, full term, spontaneous delivery; the birth weight was 8½ pounds. The patient was considered normal at birth. On the 4th day of life, a circumcision was done and at this time a mass was felt in the left upper quadrant. The consulting pediatrician saw the patient at this time and his report was as follows:

"Color is good, patient is afebrile and gaining weight. He is voiding well and has had no evident genito-urinary discomfort. On physical examination a smooth movable lemon sized mass is felt in the left upper quadrant. The bladder is not distended."

A gastro-intestinal series was done and found to be normal. On the 15th day of life, the patient was transferred to Children's Hospital with the admission diagnosis of Wilm's tumor. Family history was non-contributary.

Physical examination on admission revealed a 15 day old white male not acutely ill and appearing well nourished. Temperature was normal. Positive findings were limited to the abdomen which was soft and distended. A mass was felt in the left flank which was soft, smooth, movable and filled the entire left side of the abdomen.

Laboratory examination of the urine and stool found them both to be within normal limits. On the day of admission May 14 a flat plate of the abdomen and an intravenous pyelogram were done. The reports were as follows:

"Flat plate of the abdomen reveals the presence of a space occupying mass in the left flank which displaces the stomach upward and the intestine to the right. The mass is about the size of an orange. At the end of 4 minutes the intravenous pyelogram reveals a few flecks of radio-opaque substances on the right and at the end of 10 minutes the calices and pelvis on the right are faintly outlined and appear normal. There is a small amount of dye which on the left appears to push upwards and may represent a distorted left pelvis. At the end of 20 minutes there is a moderate amount of dye in the bladder and the left kidney cannot be visualized. At the end or 30 minutes the right kidney is emptied and the left kidney reveals a displaced pelvis upwards and medially; neither the calices nor the pelvis are well outlined. The bladder is emptied and appears normal and the left kidney is still not visualized." The interpretation was that of a

space occupying mass in the region of the left lower lobe of the kidney with the above mentioned distortion which possibly represented a Wilm's tumor.

On the day following admission a surgical consultation was obtained. The findings were as follows: "A smooth, regular outlined, ovoid mass is palpated in the left side of the abdomen which seems most likely to be a renal mass. Other structures to be considered are spleen, liver, pancreas and a retro-peritoneal mass."

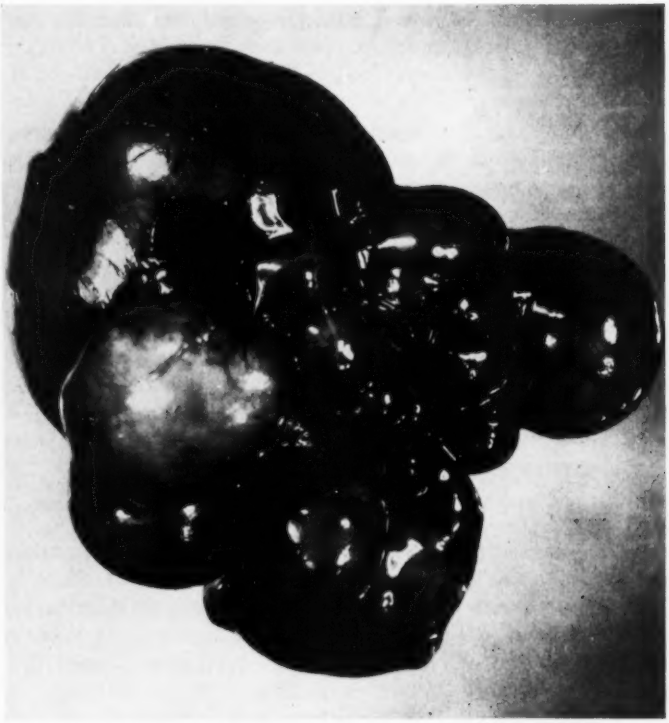

FIG. 1. SURGICAL SPECIMEN SHOWING THE CONGENITAL MULTILOCULATED RENAL CYST

On the 5th hospital day an exploratory operation was performed by Dr. Charles Howze. Behind the posterior peritoneum was a multiple cystic mass extending from under the diaphragm to the pelvic brim. The posterior peritoneum was incised over the cystic tumor. The tumor consisted of multiple cysts varying in size from that of a golf ball to a small grape. No renal tissue was apparent between any of the cysts. The mass was freed from surrounding tissue and found to be attached to a

normal sized ureter. On the lateral border of the cystic mass was found the left testicle which was removed along with the cystic tumor after division and ligation of the ureter. The right kidney was palpated and felt normal in size with no cysts on the surface.

The patient was returned to the ward in good condition. His temperature was elevated to 101° and returned to normal over a period of 24 hours where it remaind for his entire hospital stay. The patient was gradually restored to normal feeding; his post-operative course was satisfactory and uneventful. He was discharged from the hospital on the 20th hospital day, 15 days after operation.

PATHOLOGICAL REPORT

Frank Murphy, M. D. : Gross examination of the specimen revealed a multicystic mass measuring 10 x 7 x 6 centimeters (figure 1). The cysts were translucent and aspiration revealed clear golden fluid. The cyst walls measured about one millimeter in thickness. There was apparently no functional kidney tissue present. There was also a small testicle present.

Microscopic examination of a section of a cyst showed a connective tissue wall lined by a thin layer of cuboidal to flat epithelial cells sharply defined from sub-epithelial structures. Scattered blood vessels were also seen. The histological picture was in keeping with the diagnosis of a congenital cyst. There was no evidence of malignancy noted.

Microscopic examination of a section of the testicle revealed well differentiated, rather markedly congested, atrophic testicular tissue. There was nothing suggestive of neoplasm or any other pathological process.

DISCUSSION

Martin Randolph, M. D. : Campbell[1] regards congenital multilocular cysts of the kidney as an aggregation of solitary cysts and probably of the same etiology. Carson[2] in his discussion of congenital solitary cysts states that the cyst wall may occasionally be multiloculated in character. Hence this specimen either represents an aggregation of solitary renal cysts or a single solitary cyst with a multiloculated cyst wall.

While congenital solitary renal cysts are said to be infrequent, they are extremely rare in children. Of the 153 cases collected by Carson[2] only four were in children. Two were one year of age, one was four years old and one eight years old. The majority of cases reported were between 30 and 60 years of age, the average being 45 years. In Campbell's autopsy series in children[1] there were 5 cases, an incidence rate of 1:2400. Three were in males. Kretschmer[4] collected 48 cases from the literature and found only 4 cases in children.

Carson[2] in agreement with other reports[1, 4] believes the solitary serous

cysts of the kidney are the result of a congenital malformation. In general, three etiological views are held; that they are due to (1) embryonal rests, (2) failure of union between glomeruli and collecting tubules and (3) retention cysts secondary to constriction of tubules by fibrous tissue. Drachter[4] believes that the most frequent cause of renal cysts in childhood are due to trauma arising from bleeding into the kidney or fatty capsule.

Carson, speaking of abnormal conditions associated with congenital solitary cysts of the kidney[2] lists hypernephroma, cancer of the kidney, papillary carcinoma of the renal pelvis, horseshoe kidney, abnormalities of renal blood vessels, bifid pelvis, hydronephrosis, renal calculi and tuberculosis of the kidney. Campbell[1] states that other anomalies frequently co-exist. In his series of five cases in children, four had other urologic anomalies and in one case the anomaly was non-urologic. In the case reported here, the only co-existing anomaly was an undescended testicle on the same side as the multilocular cyst.

As in our case, symptoms are characteristically absent, although Kretchner[4] describes a case in a female infant of 3 months of age with vomiting since birth. The condition usually goes unrecognized until the cyst assumes the proportion of an abdominal tumor. Intravenous or retrograde pyelograms define the outline of the cyst.

Treatment consists in excision of the cyst. In our case total destruction of the kidney parenchyma necessitated nephrectomy.

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CLINICO-PATHOLOGICAL CONFERENCE

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By invitation:
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An eight year old colored male was admitted to the Children's Hospital on October 1st because of fever and a stiff neck of approximately twelve hours duration.

The afternoon before admission the boy had struck his head against an iron bed. That evening vomiting occurred twice and he complained of headache. The mother noted that he felt feverish. On the morning of admission the fever persisted and he complained of a stiff neck.

In the hospital the temperature was 103°. No evidence of trauma could be found. The reflexes showed suggestive neck and leg Brudzinski signs. The heart rate was rapid, the rhythm regular and a systolic murmur was heard over the precordium with maximum intensity in the third interspace to the left of the sternum. A lumbar tap revealed spinal fluid under normal pressure and having 4 cells per cu.mm. with 5 mgm. % of protein and a normal reduction of Benedict's solution. The temperature became normal within 2 days and he was discharged with a diagnosis of pharyngitis.

Two days later the patient returned to the hospital stating that he had a pain and swelling in the right ankle the preceding day which had disappeared. His fever had returned 24 hours after his discharge and he had an attack of epistaxis.

Past history reveals two previous hospital admissions at the age of 8 and 11 months because of bronchopneumonia with rickets and malnutrition as complicating factors. The patient attended the Heart Clinic at irregular intervals during his 8 years of life where the impression obtained that he had congenital heart disease with probably an inter-auricular septal defect and patent ductus arteriosus. An electro-cardiogram taken at three years of age confirmed the above impression. He had a tonsillectomy at 7 years of age, chicken pox the same year and measles at 4 years.

The birth was normal at Garfield Hospital, the weight being 6 lbs. 1 oz. The parents were living and well and there were no siblings. No familial diseases were known.

The physical examination on the last admission revealed a poorly developed and nourished colored male who appeared acutely ill. The skin was dry and hot. The head was held to the left side. The pupils were equal and reacted to light. The nose contained bloody crusts while the pharynx was negative. The lungs were clear to percussion and auscultation. The apex of the heart was in the 5th interspace with the left border 5 cm. to

the left of the sternum and 2 cm. to the right. The rate was rapid and rhythm regular. The examination of the abdomen was unrevealing. Neither liver nor spleen could be felt. Small cervical lymph nodes were palpable. The reflexes were normal.

X-ray examinations showed a possible pan-sinusitis and a chest film exhibited increased perihilar markings in both lungs fields. No evidence of fracture of the skull was demonstrated roentgenologically.

In the hospital the child was drowsy but responded to questioning. The temperature ranged between 100° and 104°, the average being approximately 102°. The pulse rate varied between 110 and 120 per minute. The tuberculin O.T. and P.P.D. were negative as was the Schick test.

LABORATORY EXAMINATIONS

Blood: Hemoglobin 12 gm. %, leucocytes 5,500 with 42% segmented forms, 37% band forms, 1% metamyelocyte and 19% lymphocytes. Two subsequent examinations revealed essentially the same results. Agglutination tests for E. typhosa, P. typhosus A and B as well as Brucella abortus and Proteus OX_{19} were negative. The erythrocyte sedimentation rate was 0 mm. in 1 hour (Wintrobe). The urine was turbid, neutral, had a specific gravity of 1.003, with no albumin, sugar acetone or blood cells present.

Because the patient complained of headache and a stiff neck and was drowsy, two lumbar punctures were done with the following results:

October 18: The fluid seemed to be under normal pressure and contained 85 cells per cu.mm., 20% being lymphocytes and 80% polymorphonuclear leucocytes. A Wassermann on this fluid was negative as was a culture. The specimen was inoculated into a guinea pig which died within 5 days. The latter showed a few white nodules on its spleen interpreted as an inflammatory reaction without evidence of tuberculosis.

October 21: Fluid was under no apparent increased pressure, there being 450 cells per cu.mm., 75% being polymorphonuclear leucocytes and 25% lymphocytes, chlorides 709 mgm. %, protein 30 mgm. % with normal reduction of Benedict's solution. Culture—no growth.

On October 19, three weeks after hospitalization, the patient became comatose and progressively went downhill. Four days later it was noted that he became rigid, a position of opisthotonos was assumed and convulsive movements of the upper extremities began. Stertorous breathing occurred and the rigidity became more marked. At this time the temperature rose to 106°. The patient remained unconscious and expired 12 hours later on October 23, twenty-two days after admission.

DISCUSSION

Dr. Bernard J. Walsh: I should like to make a few comments, one regarding the description of the murmur in the third paragraph. A systolic

murmur as described here carries little meaning without the details as to its loudness, whether or not it was harsh, rough, or musical, and in what direction it radiated, if it radiated at all. Unless a systolic murmur is described in this fashion it is hardly worth while to mention it.

It is certain that the patient attended the Heart Clinic irregularly and that he was thought to have an interauricular septal defect and a patent ductus arteriosus. It is hardly possible to make such a diagnosis since there is a certain similarity in the murmur and in the contour of the heart and great vessels in the two conditions. In addition, it is a rare combination. I personally have never made a diagnosis of these co-existing defects. It is a bit confusing, too, to have it said that the electrocardiogram taken when the patient was 3 years of age confirmed the impression of congenital heart disease. The electrocardiogram can rarely be said to give this information. Practically all patients with a patent ductus arteriosus have a normal electrocardiogram and very many of those with a interauricular septal defect have normal electrocardiograms also.

The laboratory data here are of considerable importance. A particularly important observation is conspicuous by its absence, and that is, blood culture studies. It may be that I am not supposed to have this information.

Dr. Bischoff: The record shows one blood culture taken which gave a growth of diphteroid organisms in 48 hours. These were considered to be contaminants.

Dr. Walsh: It seems odd that only one blood culture was taken, for it is apparent, I believe, that this boy must have had an infection. It is also apparent that the infection caused meningeal irritation, but the persistent finding of sugar in normal amount in the spinal fluid eliminated quite definitely I believe any direct bacterial invasion of the meninges. We must consider then that this patient had an aseptic meningitis. If one believes this to be so, then what was the primary site of the infection? Did it bear any relationship to the injury which the patient had at the start of his illness? There is nothing to indicate that he had a fractured skull or ruptured ear drums. He apparently did not have pneumonia. Subdural hematoma can be eliminated, I believe, because of the absence of xanthochromic character of the spinal fluid although something less than half the patients with a subdural hematoma do have clear spinal fluid, but in addition the spinal fluid pressure was apparently normal at all times. Brain abscess can be discarded, I believe, because of the normal spinal fluid pressure and the persistently normal spinal fluid sugar. The absence of evidence of increased intracranial pressure on neurologic examination is another aid in eliminating brain abscess and a solid brain tumor.

Could the patient have had rheumatic fever? I do not believe so since

we must attempt to explain the entire illness in one diagnosis, and I have never known rheumatic fever to cause such meningeal irritation and neurological developments as occurred here. Likewise the description of the heart disease does not fit that of rheumatic heart disease. It is rather unlikely that if rheumatic fever were the cause of the pain and swelling of the right ankle this would have been the only joint affected, or that it would have cleared so quickly.

There is one condition, I believe, that will explain this patient's illness and clinical course including the joint pain and swelling and that is subacute bacterial endocarditis. We would feel a bit more certain about this diagnoise if there was a better description of the cardiac findings, but since he did attend the Heart Clinic for several years it is safe to assume that he did have heart disease. From the description given to us of the heart, I believe he very probably had congenital heart disease with interventricular septal defect without any other lesion of the heart. I believe bacterial endocarditis vegatations will be found in the heart located on the septal defect and perhaps in the left and right ventricles which have given rise to infected emboli which in turn caused several cerebral infarcts. The location of the infarcts determine the neurological findings and the aseptic meningitis. It is probable too, it seems to me, that this patient was in this hospital long before the present-day therapy of bacterial endocarditis came into use.

PATHOLOGICAL REPORT

Dr. Rice: Externally the brain appeared edematous and on section a hemorrhagic infarct was found involving the cerebellum with microscopic evidence of septic embolism affecting the cerebral brain substance. The lungs and spleen were the sites of small emboli. The right kidney was ectopic being located at the brim of the pelvis over the bifurcation of the aorta. The organ was supplied by two vessels given off at the aortic bifurcation. An infarct was found in the lower pole of this kidney.

The pathological diagnosis was:

1. Developmental anomaly of the heart and aorta
   a. Interventricular septal defect.
   b. Anomalous origin or the aorta.
2. Vegatative endocarditis involving the mitral valve.
3. Mural thrombus, left ventricle
4. Infarction of the brain, lungs, spleen and right kidney
5. Ectopic kidney, right.